Extracranial carotid aneurysm in Behçet disease: Report of two new cases

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Extracranial carotid aneurysm due to Behçet disease is extremely rare. To our knowledge, this complication has been previously reported in only 12 cases. We report two new cases of extracranial carotid aneurysm in Behçet disease and discuss the clinical features, therapeutic modalities, and postoperative complications of these uncommon lesions. (J Vasc Surg 2006;43:627-30.)

Behçet disease (BD) is a multisystemic recurrent inflammatory disorder that was originally described as a triad of oral and genital ulcerations with uveitis.[1] Arterial involvement is the most common cause of mortality in patients with BD. Aneurysms are common among the arterial lesions and affect various arteries, but mostly the abdominal aorta. Extracranial carotid aneurysms in BD are extremely rare. We report two cases of carotid aneurysm due to BD and review all of the cases previously reported in the literature.

CASE REPORTS

Case 1. A 26-year-old man was admitted for a left-sided pulsatile cervical mass present for 2 months. He had a 3-year history of polyarthralgias, recurrent oral aphthous ulcers, and folliculitis. There was no history of trauma, surgery, or irradiation in his neck. Physical examination revealed a pulsating mass at the base of the left neck. Neurologic and ophthalmologic examination showed no abnormality. Blood tests showed an erythrocyte sedimentation rate of 100 mm/h and a C-reactive protein level of 55 mg/dL (normal, <6 mg/dL). Test results for autoimmune antibodies (serum antinuclear, antineutrophil cytoplasmic, and anticardiolipin antibodies) and infectious diseases, including syphilis, tuberculosis, and human immunodeficiency virus, were all negative. Ultrasonography and carotid angiography revealed a fusiform aneurysm of the left common carotid artery (CCA; Fig 1). The diagnosis of BD was made on the basis of history, the presence of systemic inflammatory reaction, and positive pathergy test results. (The pathergy test is a simple test in which the forearm is pricked with a small, sterile needle. The occurrence of a small red bump or pustule at the site of needle insertion constitutes a positive test.)

Surgical treatment was performed because the risk of rupture seemed high. Median sternotomy was performed along with a left laterocervical longitudinal neck incision. After control of the proximal and distal left CCA, the aneurysm was opened. Because of the friable nature of the proximal CCA, it was ligated, and reconstruction was performed with an aortocarotid bypass by using a polytetrafluoroethylene graft. There were no postoperative complications. The patient was discharged on the eighth postoperative day with prednisolone 15 mg/d and aspirin 300 mg/d. One year later, he remained symptom free, and Doppler ultrasonography documented the patency of the graft. Microscopically, the vessel wall showed foci of inflammatory exudation with lymphocytes, mononuclear cells, and neutrophils. Internal and external elastic bands were absent.

Case 2. A 41-year-old man was referred for a pulsating mass in his right anterior cervical region. He had a 5-year history of recurrent oral and genital aphthous lesions and skin lesions. On admission, his temperature was 37°C, and scars of old ulcers were evident on the scrotal skin. In the right anterior cervical region, a pulsatile mass was seen (Fig 2). There were no neurologic or ophthalmologic abnormalities at examination. Ultrasonography and computed tomography revealed a pseudoaneurysm of the right CCA originating just proximal to the carotid bifurcation (Fig 3). Laboratory test results were consistent with an inflammatory condition, with an erythrocyte sedimentation rate of 110 mm/h and a C-reactive protein level of 38 mg/dL. A pathergy test was performed to confirm the diagnosis of BD, and the results were positive.

Because of a concern of imminent rupture of the aneurysm, the patient underwent operation 2 days after his admission. A median sternotomy was performed to control the brachiocephalic artery. Reconstruction was performed with a bypass from the CCA to the internal carotid artery by using a polytetrafluoroethylene graft. The external carotid artery was reimplanted into the graft. The patient was discharged without major complications and placed on prednisolone 20 mg/d and aspirin 300 mg/d. No complications from this reconstructive procedure have occurred. Microscopic examination of the vessel wall revealed that the intima and most of the media were absent.

DISCUSSION

BD is a multisystemic disorder characterized by recurrent orogenital ulcers, ocular manifestations, and skin lesions. It frequently occurs in Mediterranean countries, the Middle East, and eastern Asia. There are no pathogno-

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Fig 1. Carotid angiography showed a fusiform aneurysm of the left common carotid artery.

Fig 2. Pulsatile mass in the right anterior cervical region.

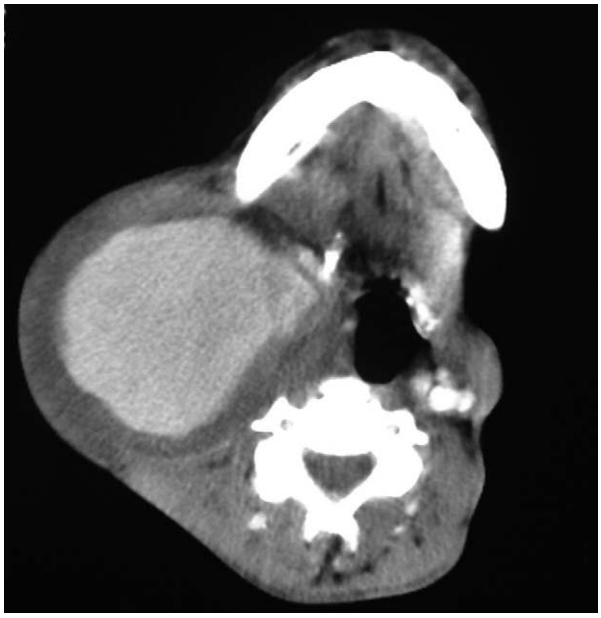

Fig 3. Computed tomographic scan of the neck showing an aneurysm of the right common carotid artery.

monic laboratory tests or histologic findings specific to BD. Thus, the diagnosis is based on clinical criteria, and various criteria have been proposed. The most commonly used criteria are those of the International Study Group for BD, and these require recurrent oral ulceration plus at least two of the following: recurrent genital ulcerations, eye lesions (such as uveitis), skin lesions (such as erythema nodosum or folliculitis), and a positive pathergy test.[2]

Vascular involvement appears in 7% to 29% of patients with BD[3] and gravely affects the course of the disease. Vascular lesions are most likely to involve the venous system; however, arterial lesions are associated with a greater risk. Arterial involvement occurs in 3% to 5% of patients, often in the form of a rapidly expanding aneurysm.[4,5] The pathogenesis of aneurysmal degeneration is thought to be vasculitis resulting in obliterative endarteritis of the vasa vasorum supplying the medium-sized and large-sized vessels.[4] Vascular involvement occurs several years after the diagnosis of BD. Nevertheless, once vascular disease develops, progression to aneurysm formation can be rapid.[5]

The most common site of aneurysm formation is the abdominal aorta, followed by the femoral and pulmonary arteries.[3,6] Aneurysms of the extracranial carotid arteries are seldom reported. In a previous article, we reported a case of an internal carotid aneurysm due to BD.[7] To our knowledge, only 12 cases of carotid aneurysms due to BD have been reported in the English medical literature (Table).[5-16] Most patients with these aneurysms were male and ranged in age from 16 to 47 years. The CCA is the most frequent location of these aneurysms, as in our cases. The chief complaint was a neck mass in most patients, but only one patient reported by Park et al[6] experienced transient ischemic attacks, presumably from thromboembolism from the aneurysm sac.

Rupture or bleeding from carotid aneurysms is rare, and there is only one case of a ruptured carotid aneurysm due to BD reported in the literature.[11] In contrast, multiple aneurysms with BD are relatively common,[10,12] and other arterial aneurysms were found in five of these patients, including aneurysms of the abdominal aorta (three cases),[5,6,10] femoral artery (one case),[12] and radial artery (one case).[12] Thus, patients with a diagnosis of BD should be screened for multiple silent aneurysms.[11] For this purpose, ultrasonography and computed tomography are more appropriate than angiography because of the risk of pseudoaneurysms at arterial puncture holes.[16]

Aneurysmal lesions in BD respond poorly to medical treatment because of an apparently increased tendency to

Table. Clinical information for 12 cases of extracranial carotid aneurysm due to Behçet disease reported in the English medical literature

Study	*Age (y)*	*Sex*	*Localization*	*Treatment*	*Long-term postoperative course*
Park[6]	25	F	Bilateral CCA	Not operated	—
Dhobb[7]	44	F	Left ICA	Resection and end-to-end anastomosis	Alive and well 1 y after operation
Suzuki[8]	16	M	Left CCA	Resection and PTFE bypass	Alive and well 2 y after operation
Tacal[9]	35	M	Right ICA	Resection and tube grafting	Carotid occlusion 2 wk later
Kuzu[5]	—	—	—	PTFE bypass	Alive
Tuzuner[10]	16	M	Right CCA	Resection and PTFE bypass	No complications at 7 y
Tuzun[11]	25	M	CCA (ruptured)	Ligation	Alive after 1 mo
Canova[12]	32	M	Right ICA	—	Alive and well 11 mo after operation
Bonnotte[13]	47	F	Right ICA	Bare stent and coil embolization	No complication 4 y after operation
Park[14]	32	M	Right CCA	Stent graft	Carotid occlusion 6 mo after operation
Özyazıcıoglu[15]	34	F	Right CCA	Resection and PTFE bypass	—
Iscan[16]	43	M	ICA	PTFE bypass	Patent graft after 35 mo
Case 1 (this study)	26	M	Left CCA	PTFE bypass	Alive and well 1 y after operation
Case 2 (this study)	41	M	Right CCA	PTFE bypass	Alive and well 11 mo after operation

CCA, Common carotid artery; *ICA,* internal carotid artery; *PTFE,* polytetrafluoroethylene; —, not mentioned.

enlarge and rupture within a short time.[3] However, although the surgical repair of these aneurysms is unavoidable, patients with BD are poor candidates for reconstructive arterial surgery because of the friable nature of the affected arteries.[3,16] Synthetic graft material is preferred to autologous vein material for repairing arterial lesions in BD because of the history of superficial thrombophlebitis.[11] Surgical intervention in the active inflammatory phase is also problematic because of the potential risks of anastomotic pseudoaneurysm formation and graft occlusion,[5] and most authors recommended avoiding surgery during the acute inflammatory phase of the disease.[16] However, in certain cases, such as the ones reported here, it is unavoidable. Carotid ligation can be used as the solution of last recourse in case of emergency and absolute impossibility of reconstruction. To potentially prevent complications from surgical repair, endovascular insertion of a stent graft has been suggested as a reasonable alternative. However, to our knowledge, only two cases of carotid aneurysms in BD treated by stent graft have been reported.[13,14] Bonnotte et al[13] reported a case of a false aneurysm in the internal carotid artery in BD that was successfully managed with a bare stent and coil embolization. In contrast, carotid occlusion occurred in a patient with two anastomotic false aneurysms treated by stent grafts.[14] Further investigation with longer-term follow-up is strongly recommended to determine the efficacy of the endovascular treatment.

Thus, we believe that an aggressive surgical approach currently represents the most suitable treatment for management of these aneurysms. Aneurysm resection and restoration of the cerebral circulation have had favorable results in most reported cases. Additionally, adjuvant immunotherapy, with or without high doses of corticosteroids, should be used to control the formation of new aneurysms and to minimize the risk of graft occlusion.[3]

CONCLUSION

Extracranial carotid aneurysms caused by BD are extremely rare. Because of their potentially high risk of rupture and cerebral ischemia, an aggressive surgical approach seems appropriate. Additionally, endovascular therapy may be an option in the management of surgically inaccessible aneurysms, but the efficiency of endovascular management remains to be demonstrated.

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